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Case Report

Primary large B-cell lymphoma involving the cerebellopontine angle: a case report

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Abstract: Primary large B-cell lymphomas involving the cerebellopontine angle (CPA) are uncommon. Fewer than 20 cases of large B-cell lymphoma at the CPA have been reported worldwide. Herein, we report a rare case of B-cell lymphoma in a 67-year-old woman who presented with dysphagia and dizziness and showed a lesion involving the right CPA on magnetic resonance imaging (MRI). The primary diagnosis was metastatic tumor; however, postoperative pathology confirmed a diffuse large B-cell lymphoma. The initial symptoms were resolved completely at the 2-month postoperative follow-up, and the postoperative course was uneventful. Large B-cell lymphoma should be included in the differential diagnosis of CPA lesions.

Keywords: CPA, swallowing dysfunction, primary central nervous system lymphoma, metastatic tumor

Introduction

Primary central nervous system lymphoma (PCNSL) is a rare malignant extranodal non-Hodgkin's lymphoma; it accounts for 0.7%-0.9% of all lymphomas and for only 0.3%-1.5% of intracranial tumors [1, 2]. Most lymphomas occur in the supratentorial areas, including the basal ganglia, thalamus, corpus callosum, and paraventricular white matter [3]. Primary large B-cell lymphomas involving the cerebellopontine angle (CPA) are extremely rare. Fewer than 20 cases of large B-cell lymphoma at the CPA have been reported worldwide [4]. Although central nervous system lymphomas are less likely to be misdiagnosed pathologically, the lack of specific clinical symptoms and imaging features make them easily misdiagnosed. Herein, we report an unusual case of a lymphoma of the right CPA and describe the distinctive clinical features.

Case report

A 67-year-old woman was hospitalized after 20 days of swallowing dysfunction and 10 days of progressive vertigo. The symptoms had begun with an unexplained cough while drinking water 20 days before hospitalization and had then deteriorated rapidly and progressed to significant coughing during both drinking and eating. In addition, symptoms of vertigo manifested as an inability to blink and nausea. Preoperative magnetic resonance imaging (MRI) showed that the lesion involved a mass effect at the right CPA. The size of the lesion was approximately 12 × 10 × 22 mm. MRI showed slightly longer T1 signals and slightly shorter T2 signals as well as homogeneous enhancement after contrast injection. Moreover, the lesion was composed of multiple separated sections (**Figure 1A-C**). Clinical neurological examination revealed no positive signs other than swallowing dysfunction. The patient had drinking profile #4 in the water swallow test. The initial diagnosis was metastatic tumor. However, dehydration treatment was provided, and the primary lesion was investigated. Whole-body computed tomography (CT) revealed no other organs with tumors, and no symptoms alleviated after medical treatment for 2 weeks. We recommended biopsy or craniotomy with pathological examination. Finally, the patient and her family choose craniotomy. No steroid treatment before or after surgery, postoperative radiotherapy, or chemotherapy was provided. Subtotal removal was seen on CT obtained 1 week after the surgery (**Figure 1D**). The symptoms of swallowing

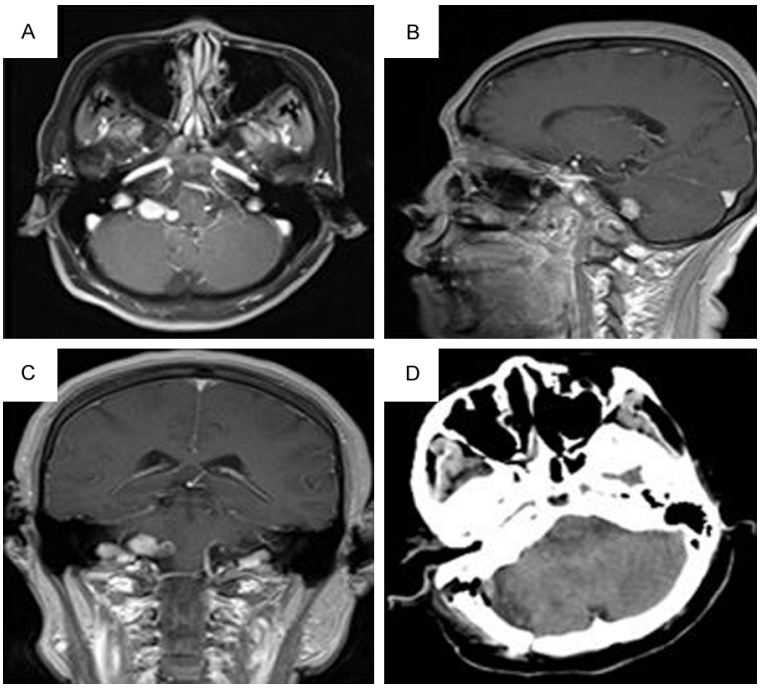

Figure 1. Preoperative axial (A), sagittal (B), and coronal (C) gadolinium-enhanced T1-weighted magnetic resonance imaging showing a homogeneously enhancing lesion in the cerebellopontine angle with compression of the medullary and bridge cerebellum. The largest section is approximately 22 × 12 × 10 mm. Postoperative computed tomography 1 week after surgery (D).

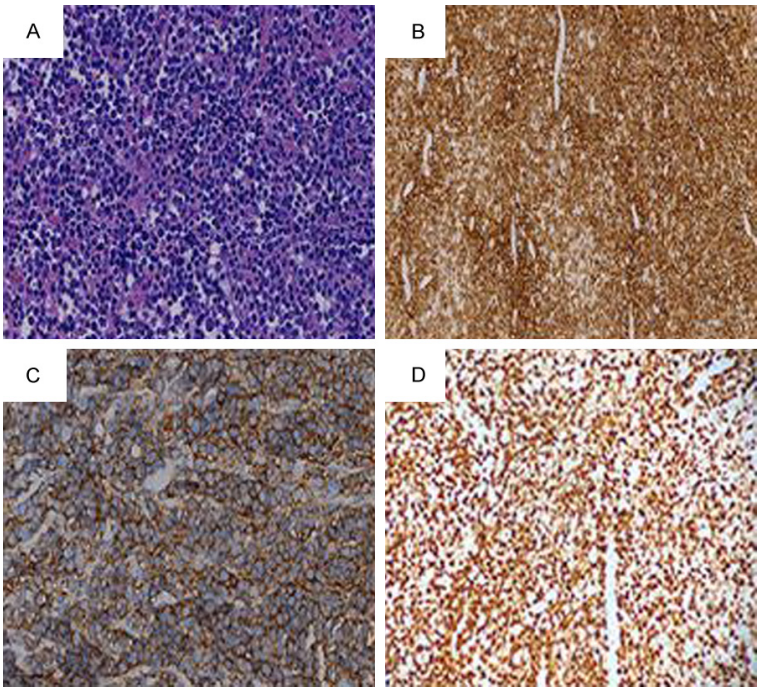

Figure 2. Histopathology (A) and immunohistochemistry (B-D) showing the tumor consisting of large, oval to round cells. Strong immunostaining for CD-79 (× 10 magnification), with membrane- (B), CD-20-membrane- (× 20 magnification) (C), and Ki-67-positive cells (× 20 magnification) (D), Over 80% expression is observed.

dysfunction and dizziness were significantly improved postoperatively. Haematoxylin and eosin (H&E) staining showed numerous closely arranged tumor cells with large and deeply stained nuclei. Immunohistochemical staining was abundantly positive for the expression of CD79 and CD20. Additionally, Ki-67 expression was increased to 80% (**Figure 2**).

Discussion

Lesions involving the intracranial CPA are mostly benign. The major differential diagnoses of a CPA mass mainly include acoustic neuroma (80%-90%), meningioma (5%-10%), and epidermoid inclusion cysts (5%-7%). PCNSL is a relatively rare, highly malignant central nervous system tumor, that accounts for only 0.3%-1.5% of intracranial tumors [1, 2]. In the literature, only approximately 20 cases of large B-cell lymphoma at the CPA have been reported worldwide. This patient was misdiagnosed at initial admission with demyelinating disease rather than a malignant tumor. Factors contributing to the misdiagnosis included the rarity of lymphomas of CPA, short disease course, and lack of unique imaging features on preoperative MRI.

PCNSL presents as a single lesion in 60% to 70% of patients, and diffuse large B-cell lymphoma accounts for the majority (90%) of cases [5], most commonly located in the hemisphere (38%), thalamic/basal ganglia (16%), corpus callosum (14%), periventricular area (12%), or cerebellum (9%) [6]. Because of these diverse tumor locations, different clinical symptoms are exhibited. The most common manifesta-

tions may be focal neurologic symptoms (approximately 70%). Neuropsychiatric symptoms are present in 43% of patients, followed by symptoms of elevated intracranial pressure, including headache, nausea, and vomiting; seizures; and ocular symptoms in 33%, 14%, and 4% of the patients, respectively [7]. According to data on primary lymphoma of the CPA collected by Vijay Seevaratnam from 1981 to 2017 [4], symptoms are mainly caused by increased intracranial pressure, auditory dysfunction, and ataxia due to VII and VIII cranial nerve injury, and the symptoms include headache, dizziness, facial numbness, hearing loss, and tinnitus. However, no reported patient with primary lymphoma of the CPA presented with swallowing dysfunction as the chief complaint. In this article, we reported a case of primary lymphoma of the right CPA in a patient with swallowing dysfunction as the chief complaint.

Central nervous system lymphomas are treated with chemotherapy, radiotherapy, and molecular targeted therapy while patients with larger masses and acute symptoms, such as cerebral hernia, can be treated surgically. In patients with newly diagnosed PCNSL, the median survival time is only 2 to 3 months if left untreated [8]. PCNSLs of CPA are rare, and most patients have been reported as isolated, rare cases. Consequently, no specific medication guidelines have been laid down. Chemotherapy with cyclophosphamide, epirubicin, vincristine, and prednisone (CHOP) or cyclophosphamide, vincristine, and prednisone, along with rituximab, a monoclonal antibody, is widely used to treat B cell lymphoma [9]. Patients who received rituximab along with CHOP reportedly had significantly higher rates of overall response compared to patients who received only the traditional regimen [10].

To the best of our knowledge, this is the first case of primary lymphoma of the CPA in a patient with the chief clinical complaint of swallowing dysfunction. Although rare, lymphomas should be considered in the differential diagnosis of CPA lesions.

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Disclosure of conflict of interest

None.

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